A Lump in the Neck

by Sara Lou O'Connor

Last summer, Dan, my always-healthy husband, went for his regular dental check-up. At his previous visit six months before, he'd met Grace, the new dental hygienist. Dan remembered he'd found it "a little creepy" when Grace had examined his neck as part of her exam. But he changed his opinion at the appointment last summer. When Grace examined Dan's neck, she felt a swollen lymph node. Our dentist also examined the swollen gland, and said it was swollen, but felt normal. He advised Dan to see his physician if the gland was still swollen in a week or so. That was June 6, 2013.

Although always-healthy, Dan had experienced a mild cold three weeks before, so we assumed his swollen gland was the result. "Soft, movable gland is normal." It says this right on the internet. The descriptions Dan read online about swollen lymph glands confirmed for him that his swollen lymph gland was normal, probably resulted from his recent cold, and should resolve itself in a few weeks.

On June 20, the gland was still swollen, so I pushed Dan to make an appointment to see his primary physician at our health center. A week later, Dan's primary doctor, an internist, agreed that the swollen lymph gland was probably nothing, but he sent Dan to an ENT for confirmation. E is for ear, N is for nose, and T is for throat. Officially, ENT doctors and surgeons are called oto (Latin for ear) laryngologists (Latin for throat). They used to be oto-rhino-laryngologists (rhino is Latin for nose), but at some point must have realized it was better to have a name more patients could say, so rhino was dropped, but the N in ENT stays. These are the specialists who know what is going on inside our ears, mouths, throats, noses, and all the connections.

During the two weeks it took to get the appointment with the ENT, we never stopped to ask why Dan was being sent to an ENT for a swollen lymph gland. We assumed because everyone thought the cause of the swollen lymph node was the cold Dan had now recovered from. People see ENTs for recurring colds and sinus problems, but we never had a discussion about it.

The ENT agreed with the internist. The gland was swollen, and it felt normal, and was probably nothing, but Dan should have an ultrasound, just to be sure.

Most ENTs are also surgeons. Our ENT performed an exam in the office, using a fiberoptic scope to look deep into Dan's mouth and throat.

The mouth and throat is called the oropharynx in ENT language, and examining the mouth and throat with a fiberoptic scope is the oropharyngeal exam. Another way to look into the throat is to put a fiberoptic scope up either of the nostrils and down the back of the throat, giving a better view of the back of the throat and bottom, or base, of the tongue. The nose and back of the throat is known as the nasopharynx, so this is called the nasopharyngeal exam.

Dan before surgery – the lump is barely visible on "left neck."

The rationale was to see if there was anything inside Dan's mouth or throat that had caused his lymph gland to swell.

The ENT saw nothing to be worried about, so we did not feel it was terribly urgent, but Dan did go ahead and schedule the ultrasound.

It took another two weeks to make an appointment that would be covered by our medical insurance.

Dan is a sound editor in the Motion Picture Industry. He has excellent medical insurance through the Motion Picture Industry Pension and Health Plan (MPIPHP). Check it out. The MPIPHP covers people in the industry, even between jobs, a wonderful system to take care of the people who work in the film business, where most of the work consists of short and long-term assignments.

Dan was unemployed at this time, due to a scheduling hole, and the insurance is designed to cover employment gaps. But with no money coming in, it was critical that we make sure that even these routine checkups would be paid for, in full if possible. We had to use only doctors and facilities approved by the Industry Network. Dan made sure to get a referral from his primary physician for each doctor and facility.

I went with Dan to the ultrasound, and the technician let me see the scan. On the screen was a lumpy ball of tissue. Even to my uninformed eye, it looked suspicious. Not normal. Large and blobby. The ultrasound technician was not allowed to comment. We had to wait until the next appointment with the ENT.

A week later, the ENT informed us that the ultrasound had shown not one but three swollen lymph nodes. He ordered a guided needle biopsy. A physician would stick a needle into Dan's neck to see what was inside the suspicious nodes.

It took several days to find another medical diagnostic center that accepted our Industry Network referral. Early on the morning of August 8, Dan and I found the building and the suite. Dan made sure they had received the Industry Network Referral and paid his co-payment.

The technician took us both into the back. I was allowed to watch as the technician positioned Dan for the surgery, locating the swollen nodes with the ultrasound. This time, as the technician pressed the ultrasound into Dan's neck, the other two nodes moved into the view on the screen.

I had to exit the room during the procedure and wait in the waiting area. The doctor would look at the ultrasound while inserting a long needle into the nodes to extract tissue. Another doctor collected the specimens during the surgery and took them to the pathology lab for examination.

Our ENT called us the following week to let us know the biopsy confirmed that the swollen nodes were cancer. Having seen the ultrasounds, I was not shocked, but I was scared.

I asked if it was a lymphoma. Lymph glands, lymphoma. I had not done much research at this time. My brother had died of leukemia almost 20 years ago. A good friend and mentor had died of lymphoma five years ago. I knew how bad lymphoma and leukemia were.

Our ENT assured me it was not lymphoma, a great relief. When we met in the ENT's office, he explained that the type of cancer was identified as squamous cell. The report from the biopsy used the initials SCC for squamous cell carcinoma. There were five additional notations from the pathology laboratory. One of these was P16+. We would learn a great deal more about P16+ later, but first, we had to deal with the mystery of the missing "primary cancer."

The swollen lymph glands mean there is cancer somewhere, probably in the mouth or throat. Our ENT performed another examination down Dan's nose into the back of his throat, but still could see nothing. He repeated the exam, but again, nothing. He had repeated the oropharyngeal and nasopharyngeal exams after the ultrasound and again after the biopsy, knowing he should be able to see the cancer, but nothing was visible that seemed irregular or suspicious.

This was now quite confusing. The SCC tissue found in the biopsy of the swollen lymph nodes had come from somewhere where there was squamous tissue. The doctor knew, from the location of Dan's swollen lymph nodes, the primary cancer should be inside Dan's mouth, nose, or throat. But nothing was visible. We had confidence in the ENT, but he could not find anything suspicious when he examined Dan's mouth and throat.

When the diagnosis had come in, we started doing research. By now we had learned that the cancer cells in Dan's swollen lymph nodes were sent there by the lymph system, but they started somewhere else. Over years of tracking thousands of patients with swollen lymph nodes related to various types and locations of cancers, cancer researchers have documented that most of the time, the location of the swollen node will indicate the specific location of the primary cancer.

Dan's swollen nodes were in the middle of the left side of his neck, spanning an area called Levels III-IV, which indicates the primary cancer should be in the oropharynx or nasopharynx.

Dan took this image in the mirror. The swollen nodes are on the left side, but appear to be on his right side. The image is reversed by the camera taking the mirror image.

Despite the mystery of finding nothing, the ENT was encouraging and said Dan had a good prognosis. The next step, he told us, was to have a PET scan, and then to see the medical oncologist who would determine the treatment.

Because nothing in medicine is 100%, the search for the primary cancer had to include most of Dan's body. PET scans (positron emission tomography) are helpful for finding and measuring cancer. They could also teach patients the importance of limiting sugar in the diet.

Before the scan, the patient is not allowed to eat any carbohydrates for 24 hours. An hour before the test, the patient gets an injection of glucose mixed with a mildly radioactive substance that lights up under the scanner. Any cancer cells in the body eat glucose. After being starved of carbs for 24 hours, cancer cells gobble the injected sugar. The radiation from the isotope in the glucose lights up, showing any suspicious areas.

Dan's swollen nodes glowed in the PET scan, but nothing else. This was mysterious, but a relief, because the primary could have been a rare one outside the mouth or throat location, or the primary could have already spread to other areas of Dan's body.

There was no sign of either, so the doctors ordered two MRIs (magnetic resonance imaging), another way to view the body to find Dan's cancer. The results were the same, no findings in Dan's brain, and none in his mouth, neck, or throat, other than his swollen lymph nodes.

By this time, we learned from continuing our own research that Dan's mysterious cancer had a name: Unknown Primary.

To be precise, the full name of Dan's cancer was: HPV-P16+ Cervical metastasis from Unknown Primary (CUP). Dan was not the only patient with SCC with swollen nodes and without a primary cancer visible in the oropharynx or nasopharynx.

We also learned more about P16+, which indicated Dan's cancer was positive for a protein marker from the human papilloma virus, usually noted as HPV.

Another medical language irony: Dan's Cervical Metastasis referred to the swollen nodes in his neck. HPV also causes cervical cancer in women, named for the uterine cervix.

This is the same virus that causes cancer of the cervix in women who've been exposed to the virus. The high rates of cervical cancer led researchers to develop a vaccination to prevent exposure to the virus from resulting in infection and eventually in cancer. But vaccinations do not help if you already have the virus and the resulting cancer.

Although at first it seemed scary that Dan's cancer was caused by a virus, we quickly learned that the P16+ was an indicator of a good prognosis. If you have an oropharyngeal cancer, it is good that it was caused by HPV. P16+ oropharyngeal cancers are much easier to treat, and patients have much better outcomes.

But another mystery. While it was good to know that HPV-P16+ was a positive sign, it did not seem possible that Dan had HPV. How did Dan get the HPV virus? Neither of us had ever had symptoms. Shouldn't one of us have had symptoms?

Most of the information we found on the internet about HPV turned out to be wrong. Only when we went to the hard-core science research sites did we find answers that made sense.

On the *Center for Disease Control* website, we learned that it is not uncommon to never have symptoms: http://www.cdc.gov/std/HPV/STDFact-HPV.htm

People may have been exposed to the virus decades before developing the cancer. We also found confirmation that a small percentage of HPV-P16+ SCCs are, like Dan's, unable to be seen by the usual cancer diagnostic tests.

Next steps...

After the biopsy had shown we were dealing with cancer, the ENT explained that he would refer us to the oncology specialists: a medical oncologist, who is in charge of chemotherapy, and a radiation oncologist, in charge of the radiation treatment.

We had assumed that surgery would be used to remove the primary cancer, but since none of the tests had revealed the primary cancer, surgery, we were told, was unlikely to find it.

The ENT could perform an exploratory surgery, using scopes and biopsies, to try to find the cancer. The procedure is known as a triple endoscopy with biopsies, because the surgeon uses three different scopes to examine the mouth, nose, and throat, all the way to the opening of the lungs. But the biopsies would be "blind," just testing tissues along the way, since nothing had shown where the primary cancer was.

We had been doing more research about unknown primary head and neck cancer. We had learned that a likely place for the HPV-OPSCC cancer to hide was in the tonsils. We asked the ENT about removing Dan's tonsils, and he agreed he could certainly do this, as part of the exploratory surgery and biopsies of the mouth, nose, and throat. Dan's insurance approved, but first we had to meet the medical oncologist.

Medical Oncology Means Chemotherapy

The medical oncologist, I'll call him Dr. MO, went over all of Dan's records and answered most of our questions. His main job, he explained, would be to prescribe the chemotherapy regimen, but he would also determine what other treatment Dan needed. We discussed options.

Our insurance covered Dr. MO as part of the Network Referral program, and would also cover the radiation oncologist he would refer us to at the Disney Cancer Center. Our other option was to go to UCLA.

Dr. MO was dismissive of what he called the "extreme surgical approach" at UCLA, which he described as disfiguring and disabling. He welcomed us to talk to the "folks" there, but said to be sure to learn how extensive reconstruction would be, and to consider the lifelong disabilities Dan would have. We assured him, we were not interested in anything that was so gruesome, if it was not necessary.

We wanted to avoid as much difficulty for Dan as possible. We asked about targeted treatment, which we'd read about on the NIH website. Was it possible for Dan to use less destructive types of medication and avoiding radiation? Dr. MO dismissed this approach as "palliative."

Dr. MO agreed that Dan could have the exploratory procedure to attempt to find the primary cancer. Whether or not the primary was found, surgery would be followed by chemotherapy, which Dan would take throughout the radiation treatment. Dr. MO explained that the chemo made the radiation treatment stronger and more effective. Together, Dr. MO assured us, chemo and radiation would obliterate any cancer in Dan's mouth, throat, or neck.

Dan's treatment would include chemotherapy using a drug called cisplatin, combined with radiation to both sides of his neck, mouth, and throat.

Dr. MO explained that he was recommending radiation treatment aimed at Dan's neck to eradicate the swollen lymph nodes, and at his mouth and throat to kill the primary cancer suspected to be there, and also to cover the other side of Dan's neck, just in case.

We would meet with the radiation oncologist after the MRI, and he would explain all this to us. The dose of radiation, 70 grays, would be sufficient to kill any cancer cells, assuring a "cure."

It seemed that Dr. MO was a good and caring oncologist. He wanted us to understand that the treatment he was recommending for Dan would "cure" his cancer. He took time to explain the steps and to help us understand the complicated names of the procedures.

Dr. MO did not feel surgery was necessary, because the chemo and radiation would kill Dan's primary cancer, wherever it was. Because exploration with biopsies was not a disabling or disfiguring surgery, he felt there was no harm in Dan having it.

Dr. MO did not discuss the probability of the primary cancer being in Dan's tonsils, and when we told him we had learned this and had asked the ENT to remove Dan's tonsils, Dr. MO made it clear he did not feel the tonsillectomy was necessary. Dr. MO usually worked with a different surgeon, but he agreed that our ENT surgeon could perform Dan's surgery.

More Learning

The ENT had explained the surgery procedure. We understood that it would involve three types of scopes to look into the different parts of the mouth, throat, nose, and top part of the lungs while Dan was anesthetized. We had also learned from our own research the reason to do this surgery. Finding the primary cancer allows the amount and area of radiation to be reduced.

By what seemed a strange coincidence, our sister-in-law's sister, Annie, had just experienced this same type of diagnostic mystery: swollen lymph nodes in her neck, SCC discovered in the biopsy, P16+ found in the pathology, but no primary cancer visible by exam or imaging. When her local oncology team recommended full-body radiation, Annie had gone to the Dana Farber Cancer Center, where her care had included surgery, followed by chemotherapy and radiation, similar to what was recommended for Dan. Annie's primary cancer was not found until the pathology of her tonsils, which were removed during the surgery.

Radiation Oncology

Next, we met with the Radiation Oncologist, Dr. RO, who works with Dr. MO. Dr. RO was very impressive. He examined Dan's nasopharynx, viewing inside Dan's throat on one screen, and looking at the images from the PET scan and MRI on another screen.

Although he could not see anything indicating a squamous cell cancer, Dr. RO pointed out to us that the doctor who had reviewed Dan's MRIs had noted a suspicious "fullness" on the side of Dan's throat opposite from the swollen nodes.

The positive nodes were on the left side of Dan's neck, and this area was inside the throat on the right side. Because of this, Dr. RO felt Dan should have full radiation from one side of the neck to the other and full radiation inside the mouth and throat.

We asked about the possibility of reducing the radiation if the primary cancer could be found in the triple endoscopy procedure. Dr. RO agreed that it might be possible, but he reminded us that very few of the unknown primary cancers are ever found.

This warning was confirmed when we found research showing that triple endoscopies with biopsies of all the possible areas only turned up about 40% of unknown primary cancers. We thought, based on Annie's results, removing Dan's tonsils would increase those odds and give him a better chance of reducing the radiation dose.

Dr. MO and Dr. RO had both met us with the good news that Dan's cancer was highly treatable. They actually said it could be "cured," with their combination of chemotherapy and radiation.

But our continuing research warned us of something else, even more troubling. Neither of the oncology specialists mentioned that 20% of the patients who use cisplatin lose a significant amount of hearing, and many more patients have minor hearing loss as a result of the platinum-based chemotoxic agents.

We asked Dr. RO about this, and reminded him about Dan's job. As a motion picture sound editor, losing his hearing would mean that Dan would lose his career and income.

Dr. RO acknowledged the problem and suggested that another type of chemo agent could be used. He held firm about the need to use chemo and to radiate both sides of Dan's neck and his mouth and throat, even if the cancer were found in the biopsies.

We next met with Dr. RO's nurse, who explained that the treatment would cause "discomfort" both on the outside of the neck and inside the mouth and throat. The dry mouth and pain might make it difficult for Dan to eat during the treatment. She gave us a list of products we could buy to help make it easier for Dan.

We already knew from hearing about Annie's treatment and long recovery that it could be very bad. Maybe by acknowledging that we knew it could be bad, we didn't get the full story.

"Quality-of-Life Implications..."

We learned more later, and not just from Annie. What both Dr. MO and Dr. RO left out was that this "treatment" severely messes up your mouth and throat, and it can cause permanent damage. They did not talk about the percentage of patients who need a feeding tube, as Annie had for months.

The radiation kills the cells in the mouth and throat, so nothing works for eating and swallowing. The radiation to the neck kills the salivary glands, so there is no moisture to protect the mouth and teeth. Annie had lost teeth, and because the radiation destroys the bones in the jaw, it is difficult or impossible to have dental work done after this treatment.

The treatment affects the taste buds so that whatever you loved eating tastes unbelievably vile. It becomes hard to get food down, so patients going through treatment get malnourished. Trouble eating persists after treatment, due to the damaged mouth and throat.

The cancer itself may be gone, but patients are left in a mess, sometimes permanently disabled by the treatment. You may not have a scar, because they do not recommend surgery to remove the swollen nodes, and they discourage any more invasive surgeries as unnecessary. You may get "cured" from the cancer, but you will be left disabled from the treatment.

Even if Dan were lucky, among the 40% of patients whose primary cancer is found in the exploratory surgery, Dr. RO felt he should still have the full radiation treatment with the chemo. Both Dr. RO and Dr. MO said the chemo makes the radiation work better.

What really happens is that the chemotoxic agent kills and damages all the cells, both the cancer cells and the healthy cells. It's easier for radiation to wipe out damaged, unhealthy cells. But even if Dan managed to survive all that and not be impaired forever, he would not be able to work as a sound editor if he no longer had excellent hearing.

The weekend before Dan's surgery, we went for a walk and talked about what he might be able to do, if even some of these impairments happened. *Teaching*? I asked. We met 30 years ago, when Dan was my Sound Teaching Assistant at USC Cinema. One of our fellow students now heads the Sound Department. *Maybe*, Dan answered, although I learned later his aspirations were not so high. He was envisioning himself at the local college's small television department. I wondered if he would lose his voice, and I knew I would miss it. As delicately as I could, I brought up the possibility.

Maybe I could be the Stephen Hawking of Sound, Dan quipped. Most of us cannot imagine a world of physical disability, yet this remarkable man gives us all the inspiration that we need to try. Using Dr. Hawking's example, Dan envisioned himself teaching with a computer, using slides instead of talking.

Our walk was a brief break in a long weekend of last-minute research. Dan's surgery was scheduled for Thursday, September 12.

The week before, I was still doing research to find something that might help. I had found articles about different diagnostic techniques, but all of our doctors had downplayed their ability to help. They were still too new and their findings could not be understood. That kind of lack of attention to our problem spurred my research.

Light in Darkness

On Friday before surgery the next week, with despair, and with my usual compulsive need to read everything, I reached the bottom of my pile of research papers. I had shoved one paper, about a new surgical technique, to the bottom assuming our trusted ENT knew best about the surgery. But nothing else I'd found had provided any help. Now I was at the bottom of the pile.

As I read about the new surgical technique in *The Laryngoscope*, the journal of the American Laryngological, Rhinological, and Otological Society, I felt like I'd found light in our darkness. The paper described using robotic arms to perform the surgery Dan was scheduled to have in less than a week. This was a new way of performing the exploration for an unknown primary. By using robotic arms and lighted, magnifying scopes, these surgeons had improved their ability to find unknown primaries, from 40% to almost 90%.

Reading this article by Dr. Mehta and colleagues, I learned that it is a very small percentage of all the squamous cell carcinomas that cannot be found through the usual diagnostic techniques. These patients, as was planned for Dan, receive chemotherapy and a heavy dose of radiation, resulting in "quality-of-life implications."

The new robotic technique allowed patients to have more directed radiation, causing less damage. I was not sure exactly what this meant, but it sounded hopeful. I spent the next few hours looking up Dr. Mehta and the other researchers who were listed as the authors on the report. None were from Los Angeles. I looked up the articles in the reference section of Dr. Mehta's article, read as many as I could find online, and then looked up those authors.

Dan was working again, but left work that Friday evening, reminding his supervisors he had to take the next two-to-three weeks off, expecting to be disabled after the exploratory surgery and tonsillectomy. He hoped he would be able to return to work briefly before his treatments started, but that was uncertain.

When Dan got home Friday night, I made him sit down and listen to the highlights of my day's reading. We both spent the rest of the weekend, except for our walk, reading medical papers written by practicing ENT surgeons and oncologists, who were researching and working with patients to discover better ways of treating the type of cancer Dan had.

The report on robotic arms had mentioned another new technique with equally positive results called transoral laser microsurgery, or TLM. I looked up the article that was referenced, read it, then found and read more articles on TLM. The corresponding author of several of the reports was Bruce Haughey (pronounced hoe-ee) from Washington University of St. Louis. I emailed Dr. Haughey to ask if there was any way he could help us with Dan's cancer. That was Saturday September 8, at 6:30 PM California time.

We had a late dinner and stopped reading long enough to confer and be sure we understood. These reports presented a very different story from everything we'd been told by our ENT and by Dr. MO and Dr. RO.

Dan had been going down this cancer path since June. Now, almost on the eve of surgery, we discover that Dr. Haughey and his team had learned how to find the primary SCC in almost 90% of the patients with unknown primaries using the TLM. The surgeons using the robotic arms had similar results. Both of these techniques could improve the chance of finding Dan's primary cancer from 40% to 90%, compared with the triple endoscopy and biopsy technique that Dan was scheduled for in five days.

Reading the statistics of patient results, what is known as morbidity and mortality, or how many patients get messed up or die, I realized that people do not die of this type of cancer, but they get very messed up and die from the treatment. Dr. Haughey's research showed that it was not necessary to kill the patient's mouth and throat to eliminate the possibility of cancer cells remaining after the primary cancer was removed. Being able to find the primary cancer made a huge difference in the patient's quality of life after treatment.

Hope for the Future

Another beacon of light, Dr. Haughey's research included patients who had not used chemo with their radiation, who still had excellent success. These patients had not had recurrences of the cancer after the initial primary cancer was removed, even though they had no radiation treatment to their mouth or throat. Only the positive node area on the outside of their neck had been treated, and that was with a reduced dose of radiation.

The next day, Sunday, I checked my email and found that Dr. Haughey had already responded to my initial query late the night before. He had sent a copy of another article I'd asked about and explained his most recent research findings. Dan printed the article so I could mark the important points.

Like the other articles, this research was amazing to us. We were in a daze. Here we were preparing to have Dan maimed forever, believing it was necessary to save his life. Now we learn about Dr. Haughey's work and that maiming may not be necessary.

Dr. Haughey could not answer all my questions in an email and wrote back to suggest we talk. I wrote back, explaining we needed to talk fast, because Dan was scheduled for surgery in four days. Dr. Haughey responded that he would have his assistant make an appointment, and he would call us the next day.

When Dr. Haughey called on Monday, September 9, he had already completed his very long day of surgery. He gave us time for a very thorough consultation, both Dan and me on the phone with Dr. Haughey. He answered all of our questions. I asked about everything, including the newest diagnostic techniques I'd discovered. He knew about them all, and he knew the answers to everything. His expertise was apparent, and his manner was kind and understanding.

I asked if he would be Dan's surgeon, and he agreed. Dan was sitting across the room using the extension phone, and his mouth fell open. I knew he was wondering how we could pay to go to St. Louis, and what if Dr. Haughey was not covered by our insurance.

Dr. Haughey had told us that he had already trained some ENT oncologists in the Los Angeles area, and Dan was thinking he could see one of them. Although I am sure they would have been good, I wanted Dan to have the best care possible, and Dr. Haughey was obviously the best doctor for Dan.

As soon as we got off the phone, we contacted our insurance company. With relief, we were told that Dr. Haughey and Washington University of St. Louis and their hospitals were all covered by our extended insurance, Anthem Blue Cross. How we would pay for the trip, we would have to figure out later.

On Tuesday, the day after our phone consultation with Dr. Haughey, after we confirmed that our health insurance would cover the costs of medical care in St. Louis, we met with our ENT in Los Angeles who was planning to do the triple endoscopy, biopsies, and tonsillectomy for Dan in two days.

We told the ENT about the research we'd done over the weekend, and showed him the articles about Dr. Haughey's work. The ENT had never heard of robotic surgeries or transoral laser microsurgery for OPSCC. He was amazed to read the results of these new minimally invasive techniques. He encouraged us to most definitely go to St. Louis and he called the local hospital to cancel Dan's surgery.

St. Louis…who knew!

A month later, Dan and I sat in Dr. Haughey's examination room. When we arrived in St. Louis the day before, we learned there was room for us at the Hope Lodge, the residence provided for cancer patients by the American Cancer Society. We were feeling very lucky.

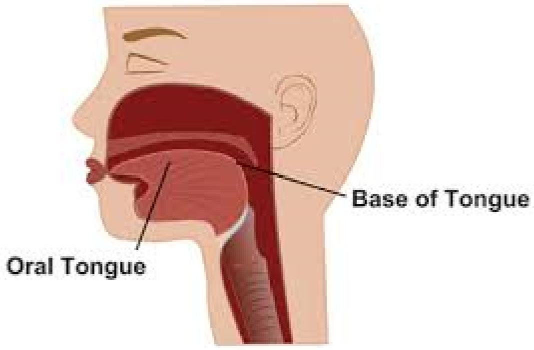
Dan in St. Louis before surgery. The lump is more visible, but still not huge.

Dr. Haughey's surgical resident had just performed another nasopharyngeal exam on Dan's throat, even though he already knew that no one had been able to find the primary cancer that should be somewhere in Dan's throat.

By then, we knew, as the resident knew, that two to three percent of the HPV P16+ cancers are not visible, or are barely visible, or are hiding under the mucosa in the base of the tongue, which is also called the lingual tonsil.

Reading Dr. Haughey's research, we had learned that Annie had been lucky to have her primary found in her upper tonsils, the palatine tonsils. Had Dan's surgery only included the palatine tonsils, as had been planned in Los Angeles, there would still have been only a 40% chance of finding the primary, because 60% of the oropharyngeal SCCs are located in the lingual tonsil, or the base of the tongue.

Dr. Haughey's plan was to remove all of Dan's tonsils. Removing all the tonsils is not a piece of cake for either the surgeon or the patient. Patients can't talk for about two weeks, and the throat is very sore, and the diet is very soft.



(http://www.keywordpicture.com/keyword/base%20of%20the%20tongue)

The tonsils are part of the lymph system that removes toxic stuff from our bodies, so it's also not great to have to lose them, but if doing so means you have a 90% chance of finding the cancer and can reduce the treatment needed for oropharyngeal SCC, it is a good trade.

Dr. Haughey's research gave him confidence that by removing all of Dan's tonsils, there was close to a 90% chance that the primary cancer would be removed, even if the pathologist did not later find the cancer in the pathology of the removed tissues.

We had not yet seen Dr. Haughey, so when he entered the exam room, it was our first meeting in person. He asked if Dan would mind another exam. Dan, always gracious, granted access yet again to his nose and throat.

As Dr. Haughey looked through his scope, he remarked, “Oh, there’s a lesion.” He looked up at us, and I’m sure my eyes were wide in shock.

This was at least the sixth or seventh time a doctor had examined Dan’s throat with a scope, and no one had ever seen anything. I said that to Dr. Haughey.

He looked again. “Yes, there it is.” He had the resident look again, and pointed out where to look until the resident saw what Dr. Haughey instantly had seen.

Just at the bottom of throat, next to the left side of the base of the tongue, just where it should be. Dr. Haughey explained that it was a “likely candidate for the primary,” although he would not know for certain until surgery and pathology.

The Procedure

Unlike the endoscopic biopsy surgery technique, Dr. Haughey’s TLM surgery includes working with a pathologist, similar to the procedure used in MOHS surgery for a skin lesion. Dr. Haughey takes a sample, then freezes a piece to be examined under the pathology scopes. The pathologist can then confirm what Dr. Haughey’s surgical scope shows him: the margins are clear. If not, Dr. Haughey takes more tissue, until the margins are clear.

Nine hours after Dan went into the surgery department, Dr. Haughey met with me to give me the excellent news. The likely lesion he had spotted was indeed the primary. The pathologist had matched the tissue to the biopsy tissue we had sent from Los Angeles the month before.

Dr. Haughey had tested the tonsils, and all were clear. There was no need to remove them. Dr. Haughey had then performed the surgery on Dan’s lymph nodes, removing the strand of nodes from Level II through IV, and confirming with the pathologist that nodes on all sides of the positive nodes were also clear: top, bottom, front, and back.

Dan woke up in the recovery room shortly after I got the news from Dr. Haughey. “When is the Dodgers game?” he asked really clearly. This raised a few eyebrows in a St. Louis hospital on the eve of the first Dodgers-Cardinals playoff game of the 2013 season.

All week, we’d smiled at the buses in St. Louis that posted “Go Cards” on their destination marquees. The nurses at Barnes Jewish Hospital of St. Louis were extremely kind to their patient, despite his being a Dodgers fan from Los Angeles.

Dan was out of the hospital in two days, not the expected four. He was eating solid food by the end of the week.

Post-surgical Treatment – Results and Decisions

Although all the surgeons and radiology professionals had interpreted the pre-surgery exams and scans as showing three positive lymph nodes, the final pathology report showed that only two were positive. But there was something else none of the scans had shown. Dr. Haughey had noted, and the pathology report confirmed, there was extracapsular spread. This meant that squamous cancer cells had moved out of the two positive nodes into the surrounding tissue.

If Dan had not had the surgery to remove the nodes, and had instead had radiation to obliterate them, we would not have known this. We would also not have known that there were clear nodes on all sides of the two positive nodes, confirmed by the pathology report.

The number of nodes found to have cancer cells is important, because it has turned out to relate to the success or failure of treatment options. The number of positive nodes is used as a guide for patients to know whether or not they should be concerned about having their cancer return. With HPV-OPSCC, only a very few cancers recur, and very few patients have a second primary appear in a year or so. Dr. Haughey has found that both – a new cancer or recurrence - have been easily removed with surgery, with no further treatment was needed. And most important, none of the patients died from their cancer returning.

Keeping track of the details of all the patients he and his colleagues have treated, Dr. Haughey has learned that the most reliable indicators of the need for additional treatment after surgery are having more than two nodes that are positive, or having a large primary cancer.

This is not unlike the prognostic findings for other types of cancer, but there are differences. Dr. Haughey's research has shown that additional treatment for patients with larger primaries and three or more positive nodes does not need to be as aggressive as what was planned for Dan in Los Angeles.

But what about the cancer cells found outside the lymph nodes? Even though extracapsular spread with other types of cancer is considered to indicate a need for more intense treatment, this has not turned out to be necessary with HPV-OPSCC.

After it was identified fully, Dan's cancer was given the official name HPV P16+ oropharyngeal squamous cell carcinoma, or in shorthand, P16+ OPSCC. Most patients who have P16+ OPSCC have the type of treatment that was planned for Dan in Los Angeles. The treatment Dan thankfully escaped is the standard of care, and it is based on chemotherapy and radiation. In the standard treatment, the primary cancer is only removed if it can be easily removed.

The doctors in charge of this standard treatment remain unaware that minimally invasive surgical techniques make it possible to remove many primary cancers with very little residual effect on the patient. They counsel patients to avoid surgery because it will be disfiguring and disabling, treating them instead with permanently disabling chemotherapy and radiation.

Most patients will take cisplatin, despite its toxic effect on hearing and other parts of the body. Both sides of the neck along with the mouth and throat get radiated with a lot of radiation. This type of radiation is not targeted to specific cells or areas, as in brain tumors, but instead covers the entire mouth, throat, teeth, jaw bone, and the skin of the neck as well as the nerves and blood vessels inside the neck, mouth, and throat.

Dr. Haughey's research shows that HPV P16+ OPSCC is different from other types of OPSCCs. The OPSCCs of the past were probably not caused by HPV, but were caused by smoking and also by excessive alcohol drinking. Those types of OPSCCs were not easy to "cure." The treatment Dan escaped was designed to try to help patients survive a large, painful, disabling type of cancer that often recurred, and which resulted in a high rate of death in a short number of years.

As we made our way through the learning and treatment of Dan's cancer, we met more and more people with this type of cancer. The CDC predicts that as many as 90% of the people in the United States have already been exposed to HPV, and the number of people with HPV-OPSCC will continue to grow to epidemic numbers. Most people diagnosed with HPV-OPSCC will go to doctors who are treating it like old OPSCC, which means a very high possibility of being maimed and even permanently disabled by the treatment.

When Dr. MO described surgery as maiming, he was describing old-style surgeries, which involved cutting through a patient's neck to reach cancer inside the throat. Before robot arms and laser microscopes were developed, it was very difficult to work in the patient's mouth. Surgeons have to keep the patient breathing, see in the dark throat, and manipulate instruments. Those old-style procedures are rarely needed now. Now, it is the standard care non-surgical treatment – the chemotherapy and excessive radiation - that is maiming and disabling.

Most ENTs and oncologists, including the medical oncologist and radiation oncologist we saw in Los Angeles, have learned that HPV-OPSCC is not as aggressive as the old OPSCC. This much of the research has broken through. But they have only learned part of the story.

For patients, the important part is that they can survive HPV-OPSCC without the heavy doses of chemo and radiation that were needed to attack and delay the recurrence of the old forms of OPSCC.

Just in the last year, the OTO - Head and Neck Surgery community have begun to discuss in meetings and in publications the results of work like Dr. Haughey's, trying to determine how to interpret these amazing results.

An editorial in the April 2014 issue of the *Journal of Clinical Oncology* describes the controversy over HPV-OPSCC. Should patients get less treatment, and if so, how do doctors know which patients will do well with less treatment?

There is a confusion of reports, made worse because most of the data do not separate HPV-OPSCC from non-HPV cancer.

An article in *Biomed Central Cancer* from 2013 laments "…management of OPSCC is now one of the most contentious and important issues in head and neck oncology practice in the wake of the growing HPV-OPSCC epidemic" (Nichols et al. BMC Cancer 2013, 13:133 http://www.biomedcentral.com/1471-2407/13/133).

What doctors call "management" would not be so difficult, nor so contentious, if they would talk with their patients, but oncologists are used to being in charge. The doctors we met, with the exception of Dr. Haughey, were good at explaining what they were planning to do and why. They were ready to answer our questions, but they did not expect us to question their plan. Their typical "management" is to lead the decision about the patient's treatment, making decisions for the patient from test results, often before meeting the patient in person.

Informed of options, with facts about possible progression of their cancer explained, patients can make their own best decisions about how to proceed.

Dan two weeks after surgery at the Botanical Garden of St. Louis

We met patients throughout Dan's journey who had made treatment decisions based on their personal beliefs about what was needed, some choosing a route of care that promised a better "cure," even though treatment effects would impair their quality of life. Others, like Dan, opted for the least invasive surgery and least damaging supportive care.

For Dan, this was laser microsurgery followed by a reduced dose of radiation and no chemotherapy. Our research gave Dan this option. Our confidence in Dr. Haughey's experience allowed Dan to minimize the destructive radiation treatment, opting for a better outcome now, with faith that if there were to be a recurrence, Dr. Haughey, or perhaps one of the surgeons he has trained can take care of it.

Doctor-Patient Relationships for Healing

Dr. Haughey is an excellent doctor and surgeon for many reasons. First, he talks to his patients, explains what is going on with them, describes their treatment options, and allows them to make their own decisions about their care. Then after Dan's surgery, Dr. Haughey described in detail the surgical results, the pathology results, and his recommendations for additional treatment.

The reduced dose of radiation Dan chose to have is called a reduced treatment plan. It is the result of many years of research with meticulous, detailed record-keeping providing the knowledge base for treatment of HPV-OPSCC at Washington University of St. Louis (WUSL).

One of the surgeons trained by Dr. Haughey was located at the Head and Neck Cancer department at the University of California, Los Angeles (UCLA). He referred Dan to Alan Chen, MD, a radiation oncologist at UCLA who knew about the treatment protocol developed by Dr. Haughey and Dr. Wade Thorstad at WUSL.

Using this plan, only the area of Dan's neck where the positive nodes had been removed received radiation. The machines that project the radiation allow gradients of the rays, so the area right over the node area received the strongest dose, then moving out from that area the gradients provided a lower dose.

No radiation was aimed inside Dan's mouth or throat, and no radiation was directed to the other side of the neck. With the reduced plan, the area with the strongest dose receive 100 times less radiation than the standard care that most patients get.

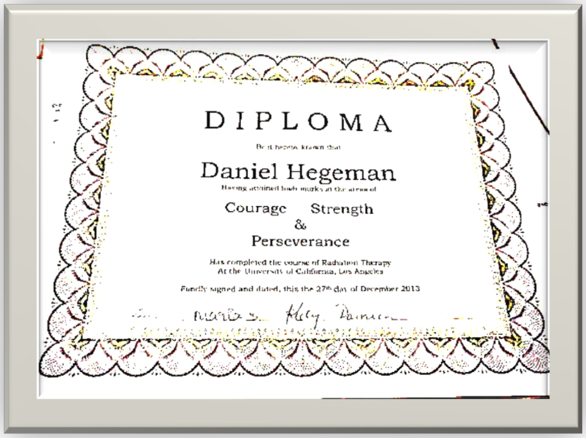
Even with this greatly reduced treatment, it was difficult for Dan to eat the last month of his treatment. He lost another 15 pounds, added to the 20 pounds he took off after he learned he had cancer and changed his diet, eliminating sugar and other carbohydrates. By the end of December, 2013, he was pretty thin.

The last weeks of the 6-week reduced dose of radiation. The scar is from the surgical node removal.

A Year Later – October 2014

Dan is preparing now for his one-year follow-up visit with Dr. Haughey. His six-month check-ups, with Dr. Haughey and with the radiation team, were both "excellent." Except for having lost weight, Dan has had very little, if any, noticeable lasting effects from his cancer experience. He returned to work within two weeks of completing his radiation treatment, and has been working all year. His hearing is excellent. His overall health is excellent.

Dr. Haughey reports that his new data will help us feel even better about the future, as results continue to show the reduced treatment plan is completely effective for treating HPV-OPSCC.



DIPLOMA

Be it hereby known that

Daniel Hegeman

Having attained high marks in the areas of

Courage Strength
&
Perseverance

Has completed the course of Radiation Therapy
At the University of California, Los Angeles

Fondly signed and dated, this the 27th day of December 2013

Dan's Dipoloma from the Radiology Team at UCLA

Two Years Later

Dan had his two-year radiology check-up in March, 2016. All is well. He continues to get to live his life as a normal, healthy man.

Three Years Later – October 2016

This month makes three years since we left St. Louis after Dan's surgery. He gets stronger each year, has gained back all his pre-illness-health diet weight, and eats too many carbs again!

This is the kind of "cure" patients and their doctors can feel good about.

Notes – January 2017

Dr. Bruce Haughey is now the director of the Head and Neck Surgery Center, Celebration Health Florida Hospital, located near Orlando in Celebration, Florida.

Dr. Allen Chen is now the Chair of the Department of Radiation Oncology at the University of Kansas, Lawrence, Kansas.

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